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Diagnosis and management of intradiaphragmatic extralobar pulmonary sequestration: a report of 11 cases☆,☆☆

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ABSTRACT

Evaluate the diagnosis and management of intradiaphragmatic extralobar pulmonary sequestration (IDEPS). We retrospectively reviewed cases of bronchopulmonary sequestrations (BPS) diagnosed in our hospital from March 2011 to May 2014, in order to identify patients with IDEPS. Diagnosis of IDEPS was confirmed using prenatal Doppler ultrasound, postnatal intravascular enhanced computed tomography, and surgery. The 11 cases diagnosed with IDEPSs were confirmed with histopathology. In our first case we did not find any mass from abdominal surgery; we then turned to transthoracic surgery. Three patients underwent thoracoscopy, and seven underwent thoracotomy. IDEPS is better approached through the chest. Thoracoscopy in experienced hands a favorable approach.

Bronchopulmonary sequestration (BPS), also known as a pulmonary sequestration, is characterized by the presence of a non-functional lung segment that receives its blood supply from the systemic circulation and lacks a normal connection with the tracheobronchial tree [1].

Two types of BPS have been described according to their anatomical and specific blood drainage features: intralobar and extralobar BPS. One of the most important anatomical differences is that in the intralobar BPS, the non-functional lung segment shares the pleura with the functional part of the lung, while in the extralobar BPS the non-functional lung segment has its own pleura. In addition, an extralobar BPS can occur in the chest, within the diaphragm, or below the diaphragm.

From March 2011 to May 2013, we previously published a series which extended from March 2011 to May 2013, during which time we identified 45 cases of BPS, 12 of them intralobar (26%), and 33 extralobar (73%). Of the latter, six cases (18%) were intradiaphragmatic extralobar pulmonary sequestration, and presented a mass within the diaphragm [2]. This rare type of BPS was first reported by Caulet in 1962 [3].

In this study we assessed a large series of cases diagnosed with IDEPS at the Women and Children Hospital at Guangzhou Medical College (China), and evaluated the diagnosis and surgical treatment associated with this condition.

1. Methods

1.1. Case selection

We selected the 11 BPS patients (8 male and 3 female) that were diagnosed specifically with IDEPS. All cases were discovered first in prenatal ultrasounds that showed the presence of a cystic-solid hyperechoic mass above or below the diaphragm. Doppler ultrasound was used to determine that the mass received its blood supply from the systemic circulation. All the patients underwent regular ultrasound checkups after birth to monitor for changes in the mass. The diagnosis was confirmed after birth using enhanced computed tomography (CT).

1.2. Surgical procedures

Our first case did not present any mass during abdominal surgery, thus we turned to transthoracic surgery. Of the ten remaining cases, seven underwent traditional transthoracic surgery, and three cases underwent thoracoscopic surgery. All surgeries were performed in our hospital by the same surgeon. After the first case, we chose thoracic surgery for all the remaining cases, either transthoracic or thoracoscopic surgery, which were all successfully completed. Operations were performed using routine tracheal intubation, and intravenous anesthesia.

In transthoracic surgery, patients lie down on the contralateral side to the locations of the mass. The diaphragm was opened using electrocauterization around the lesion, and then the mass was resected.

Thoracoscopic surgery was performed using a Stryker laparoscopic instrument with the monitors placed on the far side of the operating table, while the chief surgeon was located at the head of the operating table. We located the subscapular sixth intercostal striae and performed a small incision of about 5 mm. The incision was punctured using a 5 mm trocar. The pressure of CO2 in the pneumoperitoneum was

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Table 1
Cases detail.

Case #	Gender	Gestational age	Ultrasonic features	Position	Method	Operative Time (min)	Operation age (months)	Size (cm^2)	Complications
1	Female	24	Hyperechoic	Right	Transthoracic	180	18	2.1 × 2.2	None
2	Male	24	Hyperechoic	Left	Transthoracic	110	0.1	2.2 × 2.4	Pleural effusion
3	Female	24	Hyperechoic	Left	Transthoracic	100	7	2.0 × 2.4	Pneumonia
4	Female	22	Mixed cystic-solid lesion	Left	Transthoracic	80	3	1.2 × 1.7	Abdominal distension
5	Male	23	Mixed cystic-solid lesion	Left	Transthoracic	90	0.1	3.3 × 3.0	None
6	Male	23	Mixed cystic-solid lesion	Left	Transthoracic	90	3	1.5 × 1.7	Subcutaneous emphysema
7	Male	28	Hyperechoic	Left	Transthoracic	95	3	2.0 × 2.2	None
8	Male	25	Hyperechoic	Left	Thoracoscopic	110	0.1	2.2 × 2.4	None
9	Male	23	Hyperechoic	Left	Transthoracic	90	2	1.7 × 2.0	None
10	Male	26	Hyperechoic	Left	Thoracoscopic	80	2	2.0 × 2.3	None
11	Male	24	Hyperechoic	Left	Thoracoscopic	90	6	2.2 × 2.5	None

maintained at 4–6 mm Hg to produce an artificial pneumothorax. During artificial pneumothorax, a grasping forceps was used to oppress the lower lobe exposing the diaphragm and the costophrenic angle; this allowed us to see part of the protrusion of the diaphragm. We used electric coagulation at the diaphragm surface to isolate the lung tissue within the diaphragm. The roots of the blood supply were ligated using 4-0 Prolene suture. After resecting the mass, it was necessary to check the diaphragm for penetration towards the abdominal cavity, paying attention to possible damage to gastrointestinal and other abdominal organs. After the inspection was completed, the diaphragm was sutured using 5-0 Prolene suture to finish the surgery.

2. Results

We analyzed 11 cases of IDEPS (8 male; 3 female) that were diagnosed in our hospital from March 2011 to May 2014. The average time of prenatal diagnoses was 24.2 ± 5.2 weeks of gestation. The age at which surgery was performed ranged from three days to 18 months, with an average of four months. One patient (9.1%) presented a mass in the right diaphragm, while the other 10 patients (90.9%) presented a mass in the left diaphragm (Table 1).

In our first case, we did not find any mass during abdominal surgery, thus we turned to transthoracic surgery. Seven patients underwent traditional transthoracic surgery, while the other three patients underwent thoracoscopic surgery. The average duration of the surgical procedure was 101.4 ± 28.0 min, and the average amount of bleeding was 18.2 ± 11.9 mL. Two patients treated with thoracoscopic surgery and five treated with thoracic surgery did not need an intrathoracic drain, while the other four patients did. The average time to remove the thoracic draining tube was 2.5 days. None of the cases needed mechanical ventilation after surgery, and all patients recovered from surgery without complications. The histopathological analysis of samples taken during surgery showed the presence of bronchial and alveolar tissue covered with mesothelial cells on its surface.

Postoperative checkups were performed from two months to three years after surgery. These follow-ups assessed basic respiratory conditions, such as daily breathing, development of the thorax, check for the presence of residual tissue of the mass in the thorax using ultrasonography, and chest X-ray examinations for the detection of diaphragmatic hernias or eventrations. All the results from these checkups were normal.

3. Discussion

In the embryo, pulmonary sequestration originates from anomalies of foregut budding in which pulmonary tissues from the bronchial tree (bronchial sequestration) and the pulmonary arterial circulation, are sequestered [4–6]. Pulmonary sequestration is a rare congenital disease, and is the result of the imbalance between proliferation and apoptosis [7,8]. Two types of BPS have been described according to their anatomical and specific blood drainage features: intralobar and extralobar BPS. The extralobar type is the rarest and can be further subdivided depending on the location of the non-functional lung mass, which can be located intrathoracically or outside the thorax.

In our study, extralobar BPS was the most common type identified, occurring in approximately 73% of the cases examined. Intralobar BPS

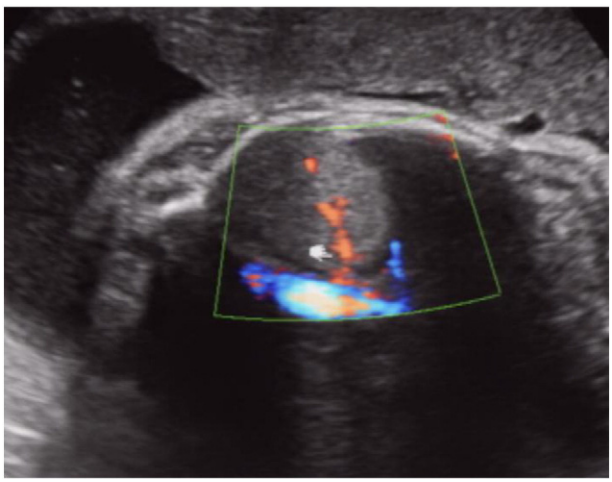

Fig. 1. Ultrasound image shows a solid well-defined mass with homogeneous hyperecho (52 mm × 32 mm × 39 mm) located between the diaphragm and the left kidney. Color Doppler shows a feeding artery from the abdominal aorta, and displacement of the left adrenal gland.

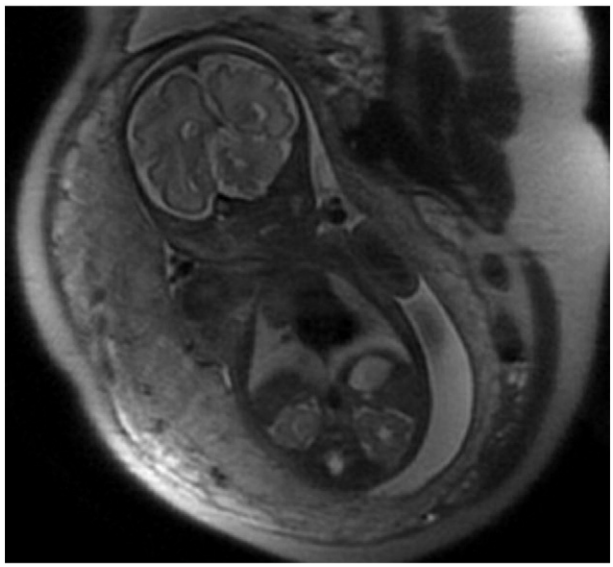

Fig. 2. Fetal MRI demonstrates the presence of hyperintensity on the left side of the chest on T1WI and T2WI images. Note that the left lower lobe was compressed.